REPORT OF A CASE OF ADIPOSIS DOLOROSA, SHOWING IMPERFECT DEVELOPMENT OF THE RIBS AND VERTEBRÆ[1]

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M. K., female, age 28 years, white, Hungarian; occupation, candy wrapper; applied at the Orthopedic Dispensary of Jefferson Hospital on May 21, 1908, complaining of pain in the back and hips. The personal and family history will be here omitted as it will be fully presented in the neurological report. At this time, she had on a spinal brace which she had been wearing for about a year and a half. Examination of the patient's back disclosed a marked high dorsal kyphosis with the apex at about the level of the fourth dorsal vertebra; a left lateral curvature most marked at about the region of the eighth dorsal vertebra. While there was no real ankylosis, the column was markedly rigid, a condition possibly due to the long-continued fixation of the spine by means of the brace. Further inspection of the other bones and joints revealed nothing of note. An X-ray was taken by Dr. Willis F. Manges, of the back including the entire spine and the pelvis, and here we had revealed an explanation of the distortion as noted above. We find the cervical vertebræ large and as well developed as we would expect of a woman of this patient's build. When we come to examine the dorsal vertebræ we find quite a marked difference, not as to the distortion alone, but we find a diminution in size both in the horizontal and vertical contours of the bodies of the vertebræ, and, as far as can be made out, of the processes as well. The ribs, while also reduced in size, have suffered the alterations in diameter only, as they are of about normal length,

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[1] Read before the Philadelphia Neurological Society, October, 1908.

the patient presenting a fair chest measurement. The X-ray of the lumbar spine and pelvis revealed no abnormalities, as they, like the cervical vertebræ, were much larger and in every way approximately normal. It is reasonable to suppose that the distortions present were due to the mechanical changes following the inequality in the development of the spinal segments, just as Bohm has called attention to the fact that numerical variations in the vertebral column apparently play a part in causing scoliosis. As to the cause of this apparent localized arrest of bony development, we may gather from the literature that the theory regarding the pituitary body as the seat of control of osseous development is fairly well substantiated. The post-mortem examinations that have been made in cases of giantism and acromegaly, seem to show that here the pituitary body is usually enlarged, while in dwarfism and cretinism, it is as markedly diminished in size. Hutchinson says, "If I were to be asked to locate a 'growth center' for the entire body upon embryonic and morphological grounds, I should unhesitatingly select the region of the pituitary body as the site of such a center."

Mr. Hastings Gilford, at the first meeting of the Royal Medical and Chirurgical Society, read a paper on a condition of mixed premature and immature development, the patient a youth of eighteen years whose skull was as yet undifferentiated into two tables and was of a thinness and appearance of a child of two months old; the scapulæ were unusually small while the clavicles were so tiny as to remind one of the "merry thought" of a chicken. On the other hand, the lower ends of the humeri and of the femoræ and knuckles of the fingers and toes were relatively large and conspicuous. While this case was one of dwarfism, it illustrates marked inequality in the degree of growth of the several parts. He called the condition micromegaly and divided the cases into two primary groups: (1) micromegaly affecting the whole body, (2) micromegaly affecting isolated structures. The latter grouping suggests such a case as ours. He again writes of a condition, Ateleiosis, and defines it as a disease characterized by a conspicuous delay of growth and development, and says that like cretinism, it may appear either before birth, during infancy or early childhood. In all likelihood, the disease as a rule becomes so ill defined, when it appears in these later years, that it can only be recognized as a mere eccentricity of normal development or is lost altogether in those variations to which all life is subject.

Osler, in his text book, under the heading of acromegaly quotes the analysis by Furnival of the recorded autopsies, 34 in number. Changes in the pituitary gland were found in all and in the majority there were hypertrophy or tumor. The hypertrophy of the gland in the majority of cases was confined to the anterior or the glandular portion of the body, and pathologically is a true hypertrophy. Hutchinson cites 12 cases of tumor formed of the gland in which no acromegalic symptoms were present. During the examination of the patient as recorded, it became apparent that the conditions presented an additional symptom group that suggested adiposis dolorosa. The case was then referred to the Neurological Department.

Neurological Examination, May 22, 1908. The family history is as follows: Her father died from injuries due to a fall. Her mother is living and well. She has one sister living and well: one sister died of diphtheria and one of pneumonia; one brother died of appendicitis, one in a "spasm" and a third when fourteen days old. Her previous history presents the usual diseases of childhood. She has suffered with headaches since her ninth or tenth year, being unrelieved by glasses. Menstruation commenced in her eleventh year, has been regular, and not especially painful. During her early menstrual history, she would frequently flow for two weeks, but in the past few years, she has passed to the other extreme, menstruation lasting about one day. She married when seventeen years of age, but has never been pregnant. Her husband is living, "well and strong." Seven years ago, she had a dilatation and curettement of the womb for menorrhagia, and two years ago was operated upon for "tumor of the left ovary." She has never had any serious illness, and uses alcohol very moderately. No history of syphilis could be obtained.

For the past eleven years, she has had pain in her back and left hip, accompanied by a certain amount of deformity due to spinal curvature. During the last year and a half she has been wearing a spinal brace, but the pain was unrelieved.

For the past two years, she noticed that her flesh was very painful, particularly about the back and knees; this pain was both spontaneous and induced by pressure or manipulation of the flesh.

She noticed that during the past year, she would often have "black and blue spots," appearing without any apparent cause. Paresthesias (crawling sensation and numbness) have at times been present, usually over the left side and thigh. There has been an increase in her weight from 105 pounds seven years ago, to 166 pounds, her present weight.

Asthenia has been present, but not marked. To use her own words "I feel so tired since I got so fat." Vomiting has occurred at intervals for the past three years. She complains of dyspnea and tachycardia but perspires freely. She denies flushing.

Upon examination, the patient was found to be moderately obese, with circumscribed masses of fat about the shoulders and knees, and upon each side of the back just below the scapulæ. These masses were extremely painful upon manipulation. There was present a moderate kyphosis.

The gait and station were normal, the pupils equal and reacting freely to light and accommodation. There was no paralysis nor muscular atrophy, but the general muscular strength as tested by resistance, grip, etc., was distinctly weak. The reflexes of the upper extremities were apparently normal, while the knee jerks were distinctly exaggerated. She presented no tremors. Sensation was normal or slightly diminished to touch, pain and temperature changes. No mental abnormality was observed beyond a mild hypochondrical tendency and a proneness to react to suggestion.

In consideration of the acknowledged role played by the pituitary as influencing bony changes and the frequency with which pituitary changes have been found in adiposis dolorosa, we believe this case to be of interest as showing evidence of arrested bony development in a patient presenting the typical symptoms of adiposis dolorosa. The bony condition, it would appear, is due to some disturbance in the function of the center regulating skeletal growth, possibly the pituitary body. And may not some later change in function of this same gland or the thyroid, be responsible for the syndrome of adiposis dolorosa? This of course can be but a matter of conjecture but one we believe worthy of further consideration. As to treatment, the patient has been conjointly treated in both departments receiving graded exercise and thyroid extract with the result that she can dispense with the brace comfortably and complains less of the pains and weakness.